A HISTORY

OF THE

CHRONIC DEGENERATIVE DISEASES

OF THE

CENTRAL NERVOUS SYSTEM.

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GLASGOW:
ALEX. MACDOUGALL, 68 MITCHELL STREET.
1895.

GLASGOW:
PRINTED BY ALEX. MACDOUGALL.

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INTRODUCTION.

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THE essays included in this work formed part of a Graduation Thesis entitled "Essays in Neurology: Historical and Clinical," and are now reprinted, with but slight alterations, from the *Glasgow Medical Journal* for 1895. They are historical studies of those chronic disorders which depend upon primary degenerative changes in the structure of the central nervous system. It is still doubtful whether the peroneal type of family amyotrophy ought to be admitted to this list, but its history is not lengthy, and I have thought it best to include it in the series. On the other hand, I have decided to leave out of consideration in this place two diseases—primary optic atrophy and general paralysis of the insane—which might, from many points of view, be regarded as belonging to this group.

Each essay is introduced by a list of designations which have been applied to the disease whose history it narrates.

The references collected at the end of each essay are intended to indicate only such treatises and articles as I have myself consulted in the preparation of the essay; and for the constant courtesy and kindness with which he has assisted me to get access to this large body of literature, my hearty thanks are due to Mr. Alex. Duncan, Librarian to the Faculty of Physicians and Surgeons of Glasgow.

Any value that may be possessed by these historical essays is, to my mind, much enhanced by Professor Gairdner's comments and criticisms which I have introduced in the form of footnotes. When Dr. Gairdner was examining my Thesis, he took the trouble to write down in that volume some observations which occurred to him, thinking that it might interest me to read them when the Thesis again came into my hands. I need scarcely say that I am very grateful indeed for these notes, and my thanks are due in addition to Dr. Gairdner for his kind permission to reproduce them here.

T. K. M.

December, 1895.

I.

HISTORY OF TABES.

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I.

HISTORY OF TABES.

NOMENCLATURE.

Progressive locomotor ataxia (*Duchenne*).
Progressive locomotor asynergia (*Trousseau*).
Posterior spinal sclerosis.
Inflammation or sclerosis of the posterior root-zones of the spinal cord (*Hammond*).
Ataxic tabes dorsalis (*Charcot*—by way of contrast to what he named spasmodic tabes dorsalis).
Tabes dorsalis (*Romberg*), or T. dorsualis.
Consumption of the spinal cord.
Grey degeneration of the posterior columns of the spinal cord (*Leyden*).
Duchenne's paralysis, or Duchenne's disease (*Trousseau*).
Tabes dorsalis, T. cervicalis, T. cerebellaris, &c. (*Remak*—different adjectives being employed according to the predominance of different symptoms).
Posterior leucomyelitis (*Vulpian*).
Myelophthisis.
Sclerosis of the posterior columns (*Erb*).
Progressive spinal paralysis (*Wunderlich*).

HISTORY.

Priority in the recognition of this disease is claimed by an English and a German writer, each for a countryman of his own; whilst in France, a tendency is shown to give a Frenchman a position of at least equal merit with observers of other lands. Thus, Dr. Gowers says: "The first really exact account of the disease was published in 1847 by Todd" . . . "The credit of the discovery of the disease belongs, if to anyone, unquestionably to Todd, and few diseases can, with greater truth, be said to have been 'discovered.'"[1]

Professor Leyden says: "Das Krankheitsbild der Tabes dorsualis ist unstreitig zuerst in Deutschland erkannt und präcisirt worden. Romberg, in seinem Lehrbuche der Nervenkrankheiten, hat die erste classische Schilderung dieser Krankheit gegeben."[2]

And M. Raymond says: "Deux hommes out surtout contribué à donner à cette maladie son individualité clinique, Romberg et Duchenne."[3]

When, therefore, a matter of this kind is in such dispute, it is our duty to go back to original sources in an enquiry after historical accuracy.

The expression "tabes dorsalis" was employed by the Hippocratic writers, say 400 years before the Christian era, to indicate a form of marasmus due to sexual excesses, which led on to what we should nowadays term "phthisis," or to some other allied disease, but which, at the same time, presented certain resemblances to diseases of the spinal cord. This was the sense in which the name was employed from the time of Hippocrates[4] to the third decade, at least, of the present century. Thus it comes about that, in recent centuries, we have had an abundance of literature dealing with groups of symptoms which a modern diagnostician would refer to under one or more of the headings, "dyspepsia," "neurasthenia," "sexual hypochondriasis."

History tells us that the famous Greek physician, Democrates, of Abdera, found the tabes dorsalis "so destructive to

our brave youth of Abdera," that he felt himself impelled to study its causes. With a view to this, he dissected the generative organs of animals, and he devoted himself so earnestly to this research that his fellow-townsmen, by whom he was much beloved, became alarmed for his health, and sent for Hippocrates, who was then residing at the court of Perdiccas, King of Macedonia. They thought their physician had gone mad, and they believed that, if they lost him, their country would be ruined. Hippocrates, however, was overjoyed when, on his arrival, he learned the nature of his professional brother's pursuits, and, after a few days' residence in Abdera, was escorted back to Macedonia in safety.

Many of the most distinguished physicians of last century wrote on this subject. Thus, Tulpius, the burgomaster and physician of Amsterdam, in explaining the consequences of certain sexual vices, says: "the spinal marrow does not only waste, but the body and mind both equally languish, and the man perishes a miserable victim."

Boerhaave, of Leyden, says that such vice causes a "dorsal consumption," and he refers to a case of this kind in which a young man "became so deformed before his death, that the fleshy substance which appears above the spinal apophyses of the loins was entirely wasted."

Mieg, of Basle, to whom, it is said, his country was indebted for the discovery of inoculation; Van Swieten, of Vienna; Hoffman (or Hoffmann), one of the most celebrated practitioners of last century, who distinguished himself by curing Charles VI and Frederick I of inveterate diseases after other physicians had failed; and Ludwig — all taught similar doctrines.

In 1800, H. St. J. Neale published, in London, a work of considerable importance, entitled "Practical Essays and Remarks on that Species of Consumption Incident to Youth, and the Different Stages of Life, commonly called Tabes Dorsalis; with an Account of the Nature, Causes, and Cure of that Distemper and the Diseases arising therefrom, especially the Nervous Atrophia and the Phthisis, or Consumption in General."[5]

Neale remarks that in such cases (viz., spermatorrhœa) either nervous consumption or consumption of the lungs may put an end to life, but there is one noteworthy distinction between the two classes, for in the nervous variety the patient is despondent, while in the pulmonary form he is hopeful. He says that eunuchs are not liable to nervous consumption.

Works of this kind seem to have appealed to a large circle of readers, inasmuch as one of them, a "practical" essay by J. Smyth (in the title of which tabes dorsalis and the venereal disease are associated) had reached its twenty-sixth edition (London) in 1792; while another, "by a physician of Bristol," reached at least six editions in London, and was published in Italian (Venice) in 1785. Yet another "Practical Essay upon the Tabes Dorsalis" (London, 1748) appeared anonymously; but, judging from some remarks in Neale's book, this was probably due to one Lewis. It contains some items of great interest. Thus, among the symptoms mentioned as characteristic of the disease in question, there are not only the regular ones of the class repeatedly alluded to, but, in addition, the "gutta serena" (*i. e.*, amaurosis) "peculiar to the tabes dorsalis," and characterised by dilation and immobility of the iris; insensibility of the optic nerve; and also, as was shown by the dissections of Bonetus and Cheselden, atrophy of the optic nerves. A digression may be here permitted, in view of certain questions which have recently been under discussion, to state that this essayist attributes the appearance of secondary sexual characters at puberty to absorption of the "succus genitalis." His remark, moreover, that "the best description of the tabes dorsalis is that of Hippocrates," helps us to understand how rapidly nervous pathology has advanced between 1748 and the present day, as compared with the period between the Hippocratic era and 1748.

The appearance of E. Smith's "Practical Observations on the Tabes Dorsalis or Nervous Atrophy, that Species of Decline induced by" sexual excesses, in 1823 (Liverpool), shows that the same notions still held their ground; so that the great advance in neurology just referred to had not yet fairly begun.

As a matter of fact, it has been made almost entirely since the completion of the first quarter of this century.

Nevertheless it is plain that, in spite of their scanty means of diagnosis, these old observers were not altogether wrong; and it is not improbable that the muscular atrophy and deformity described by Boerhaave, and the insensibility of the optic nerve described by the anonymous author, occurred in genuine cases of spinal disease.

A new era, however, in the history of tabes dorsalis began with the publication at Berlin, in 1827, of Hahn's dissertation, entitled "De Tabe Dorsuali Prælusio," in which, for the first time, the name "tabes dorsalis" (or "dorsualis") was restricted to a disease of the spinal cord, corresponding in the main, though of course only in a vague manner, with the condition which we so designate at the present day.

The next period in the history of this malady may be conveniently taken as extending up to 1851—the year of publication of the second edition of Romberg's "Lehrbuch der Nervenkrankheiten."[6] This is perhaps unfair to one of the great English observers (Todd), but unfortunately his writings seem not to have become by any means so well known as they deserved among Continental investigators. Nevertheless, an endeavour will be made to do him justice here. To show how slight is the acquaintance possessed by some well-known authorities with the work of their contemporaries and predecessors, it may be stated that Professor Leyden, writing in 1883,[7] says that Duchenne was the first who recognised inco-ordination, as distinguished from weakness, to be the prominent feature of this disease. On the other hand, Professor Erb, writing several years earlier,[8] states that Bouillaud (1845) and Todd (1847) recognised the motor disturbance to be inco-ordination and not palsy. Now, there is no doubt that Bouillaud knew the difference between ataxy and paralysis. He it was who, for the very purpose of marking this distinction, borrowed the term "ataxy" (which, like the term "tabes," comes to us from Hippocrates) from the older writers, particularly Selle and Pinel, by whom it had been applied to certain fevers, such as were called by other

authors "nervous" or "malignant" fevers. Yet there is no reason to believe, from what Bouillaud wrote about ataxy, that he ever thought of it in connection with tabes dorsalis, or, indeed, with any definite disease of the spinal cord. Todd's position, then, which will be alluded to later on, marks a great step in advance of Bouillaud's.[9]

In Germany, one of the principal writers, after Horn (1827) and before Romberg (1851), was Steinthal (1844), who gave a clinical history of the disease, and also put on record the first autopsy in Germany.

In France, the earlier investigators were Hutin,[10] Ollivier d'Angers, and Cruveilhier. About 1827, Hutin showed to the Société Anatomique, a spinal cord whose posterior columns were degenerated. (The microscope was not yet employed in researches of this kind.) Monod (1832) worked at the same subject. A glance through an essay which Ollivier (d'Angers) had published in 1824—"De la Moelle Epinière et de ses Maladies"—is enough to show that the knowlege possessed at that date of the morbid anatomy of the cord was, as nearly as it might be, *nothing*.

Cruveilhier had an excellent field for observation at the Salpêtrière, whither all the incurable paraplegics and hemiplegics of the Paris hospitals ultimately found their way. In his "Etudes sur la Paraplégie" he describes the case of a woman, aged fifty-two, who had complete loss of sensation in the lower limbs, with incomplete loss of motor power, and a gait like that of a drunk person. She often fell, and she once broke her leg in this way, it is said. The fracture caused no pain, either at the time, or during treatment. When lying in bed she could execute almost any movement. There was also some numbness and tingling in the fingers, but the power of movement in the upper limbs was almost perfect. The necropsy revealed tubercular changes in the lungs, and yellowish grey degeneration of the spinal cord, limited to the posterior columns. These were involved in their whole breadth in the lumbar and dorsal regions, but in the cervical region degeneration was confined to the tracts bordering on the median furrow. The author's remarks are of much

interest. He says that in this case paralysis of sensation was much greater than that of movement, but in other cases he has observed grey degeneration of the *posterior median* columns to affect movement much more than sensation. Therefore, he argues, one cannot conclude that the posterior median columns preside exclusively over sensation.[11]

A case such as this is clear as daylight to us, but it shows very plainly some of the riddles that had to be solved before the pathology of posterior sclerosis could be established on a secure basis.

In Germany, Jacoby, in an essay entitled *Exemplum Tabis Dorsualis* (Berlin, 1842), insisted on the localisation of the lesion in the posterior columns and nerve-roots, the anterior columns and roots being intact.

In 1844, Steinthal published a good clinical history of a case, together with the results of the *post-mortem* (made by Froriep). It was a well-marked instance of tabes with motor inco-ordination. The lesion in the cord was in its posterior portion, and there was atrophy of the posterior roots in the dorsal region and in the cauda equina; the optic nerves and chiasm and the right optic thalamus were also atrophied. The anterior nerve-roots were normal.

In Britain, in 1840, Mr. Edward Stanley read a paper to the Medico-Chirurgical Society of London on a case in which well-marked changes were found, after death, in the posterior columns, from the lower end of the cord up to the pons. Disease of the anterior columns or roots had been anticipated, but these were found to be normal. It was, therefore, inferred that the doctrine of the possession of distinct motor and sensory functions by the anterior and posterior nerve-roots respectively could not be extended *simpliciter* to the anterior and posterior columns respectively. The clinical report of the case shows that the upper limbs were normal. The lower limbs were normal as regards sensation. At first, by a great effort, the patient could, while sitting in a chair, raise his legs from the ground; but latterly he failed to do this. Mr. Stanley referred to a case recorded at Paris in 1838 by P. Malle as having occurred at Strasbourg. It was characterised by softening of

the posterior columns from the fifth cervical to the third dorsal level; during life there had been loss of mobility of the upper limbs, but not of sensibility.[12]

The next writer to be mentioned is Todd (1847), whom Dr. Gowers credits with the "discovery" of the disease. Todd, arguing against the view that the posterior columns have a sensory function, appeals to the teaching of normal and morbid anatomy, and remarks that even Sir Charles Bell had given up this view in his work published in 1844. He quotes the cases of Webster (where he had himself seen the spinal cord) and of Stanley to prove that sensation may exist in the lower limbs independently of the posterior columns. He also cites cases recorded by Cruveilhier, Budd, and Serres, where motion was lost and sensation was preserved, though the posterior columns were diseased. Todd, therefore, expresses himself as being strongly of opinion that the office of the posterior columns is something very different from anything hitherto assigned to them. They might be in part commissural between different segments of the cord, and in part subservient to the function of the cerebellum in regulating and co-ordinating the movements necessary for perfect locomotion. "In many cases in which the principal symptom has been a gradually increasing difficulty of walking, the posterior columns have been the seat of disease. Two kinds of paralysis of motion may be noticed in the lower extremities—the one consisting simply in the impairment or loss of the voluntary motion, the other distinguished by a diminution or total loss of the power of co-ordinating movements. In the latter form, while considerable voluntary power remains, the patient finds great difficulty in walking, and his gait is so tottering and uncertain that his centre of gravity is easily displaced."[13] In two cases of the latter kind, Todd predicted disease of the posterior columns in accordance with this theory; and his predictions were verified by *post-mortem* examinations. On looking over the records of cases of disease of the posterior columns, he finds that they all seem to have shown, in the first place, locomotor disturbances, sensation being

affected only when disease extended to and involved the posterior roots of spinal nerves.

The case recorded by Dr. Webster, and mentioned by Todd, was reported to the Medico-Chirurgical Society of London on the 8th November, 1842, and was afterwards published [14] as a "Case of Paralysis, without Loss of Sensation, from Disease of the Cervical Medulla." The description makes it clear that it was not a typical example of tabes. The lesion in the cord was not confined to the posterior columns. Todd examined the cord microscopically, and found that the antero-lateral columns, the posterior horns, and probably the anterior horns also, were involved. He believed it supported his theory that the posterior columns do not convey sensory impressions to the brain, or, at least, are not the only channels through which such impressions pass.

No doubt Romberg owed much to our great countryman, Sir Charles Bell, whom he styles "the Harvey of our century," but he was himself no mean authority; and the appearance of the second edition of his well-known Text-book (1851) must be regarded as marking an epoch in the progress of neurology. He recognised the disease now under consideration as a distinct morbid entity, and appropriated for it the designation "Tabes dorsalis," which, though of ancient origin, had as yet possessed but an ill-defined significance. He gives a tolerably full description of the malady; points out the symptom now known by his name—viz., the insecurity on standing erect in the dark, or with the eyes shut (he says he called attention to this ten years before); and notes the frequent occurrence of amblyopia, and—even when the optic nerve is not implicated—"a change in the pupils of one or both eyes, consisting in a contraction with loss of motion." Patients with this disease, "if they are members of the higher classes, anxiously endeavour to conceal their loss of motor power, in order to avoid the evil reputation of being affected with tabes dorsalis." This is an indication of the extent to which the idea that sexual excesses were the cause of the disease known as tabes dorsalis still lingered in the popular mind. Romberg says that the first symptom is reduction of the motor power

in the muscles, first and foremost in the lower extremities. "The patient complains of weakness and inability to perform any movements," and the sense of touch and the muscular sense are diminished early in the disease. This writer does not attribute nearly so much importance as previous authors had done to sexual excesses in the etiology of tabes; he rather thinks that rheumatism from exposure, hard work, &c., is a more fruitful cause. Romberg describes further the principal naked-eye lesions.

It is particularly to be noted that Romberg, writing four years later than Todd, failed to recognise that in this disease he had to deal with motor inco-ordination, and not with motor paralysis.[15]

In 1855, Dr. J. Russell Reynolds published his work on *The Diagnosis of Diseases of the Brain, Spinal Cord, Nerves, and their Appendages.* In this treatise, the distinction between "anæsthesia muscularis" and loss of power is clearly noted, as well as the frequent association of diplopia with the former condition. "It appears most probable (especially since this is the first change in cases which subsequently exhibit perfect paraplegia) that the centripetal tract of fibres is affected."[16] The author states that he has repeatedly found loss of the muscular sense to exist alone in cases that had been termed paraplegic, but that this condition ought to be carefully separated from paralysis in the ordinary sense of the word.[17]

Palsy of the muscular sense had been noted by Charles Bell in 1822, and Duchenne had described analogous pathological facts in 1850.[18]

In 1858, Dr. (afterwards Sir William) Gull recorded[19] a well-marked case of locomotor ataxy, with the spinal lesion limited to the posterior columns. It is included in his second series of "Cases of Paraplegia." Dr. Gull remarks that this case confirms Todd's theory as to the functions of the posterior columns.

The next period in the history of tabes is marked by the publication, in 1858-1859, of Duchenne's papers, *De l'Ataxie Locomotrice Progressive.*[20] Duchenne's account of the disease was fuller than any that had yet been published, as is

admitted even in Germany. This affection was to him absolutely new, for it was the practice of this extraordinary man (who never had an hospital appointment) to read the writings of others after he had completed his own researches, and not till then. Believing the disease to be as yet undescribed, he gave it the name which recalls its most prominent symptom. He discovered the malady for himself, but his is not the merit of priority. Nevertheless, his description was quite new to his countrymen, and a French writer has said: "Who does not recollect the astonishment exhibited in the clinic by that experiment of Duchenne of drawing from his bed a patient regarded as absolutely paraplegic, and loading him with the weight of a man of ordinary size, without his ever flinching under it?"[21] Duchenne's attention was first called to Romberg's description in September, 1863.

About 1852, Duchenne was engaged in the investigation of the force of muscular movements in health and in disease; and he soon found that many affections called "paraplegias" or "general paralyses" were no such thing: nay, that in many cases the power of movement, if actually measured, would be found to be considerable. In 1857, he made a communication to the Medical Society of Paris on this disease as a distinct morbid entity ("comme espèce morbide distincte").

As Dr. Poore remarks, Duchenne's picture of locomotor ataxy must still be regarded as almost complete, comparatively little having been added to it since it first left his hands.[22] Trousseau, by his famous *Clinical Lectures*, did a great deal to make Duchenne's observations widely known.

It now remains for us to deal with the facts that have been added to our knowledge since Duchenne wrote; a few are of enormous importance, as occurring very early in the disease, and so giving the physician a clue to the nature of the malady at a much earlier stage than could until lately have been hoped.

First, then, there is the loss of the knee-jerk, for this name is as convenient as any other of the numerous terms which have been applied to the same phenomenon. The recognition

of this symptom is due to C. Westphal. This writer had noticed, in 1871, that blows on certain tendons could call forth muscular contraction, and he continued to study these "tendon-phenomena," as he called them, in cases of disease. In his first published paper on the subject of tendon-phenomena (1875), he pointed out that the "knee-phenomenon" is abolished in tabes. This was speedily corroborated by Erb and O. Berger. In a very important communication[23] made, on the 7th November, 1877, to the Medical Society of Berlin, Westphal showed the great value of this symptom for early diagnosis. In a later article (1881),[24] he discussed the question of the seat of the lesion which abolishes the knee-jerk, in connection with a case which ended fatally at a very early stage of the spinal disease, the knee-phenomenon having been still present when the patient first came under observation.

Erb concluded that the loss of the knee-jerk was the most constant and the earliest of the symptoms of tabes; but, before very long, some exceptional cases were put on record, in which the knee-jerk persisted.

Ernst Jendrassik, of Buda-Pesth, showed, in 1883,[25] that during strong muscular activity the knee-jerk is obtainable in more marked degree than under ordinary circumstances—a fact which is taken advantage of by physicians in cases where, on first testing, the jerk appears to be diminished or absent.

In the second place, there is the pupil. That symptom which is known as the Argyll-Robertson pupil is present in a large majority of cases of tabes. It was first described by Dr. Argyll-Robertson, of Edinburgh, in 1869.[26] It is a pupil which does not contract to the stimulus of light while still contracting in accommodation, or, to speak more accurately, in convergence of the visual axes. Dr. Argyll-Robertson's observation was made on an ataxic patient who complained of dimness of vision in both eyes. Neither pupil contracted to light, but both contracted when the patient looked at a near object. Both pupils were very small, and were difficult to dilate with atropine. The visual fields were contracted,

and there was some degree of colour-blindness. Dr. Robertson considered the myosis to be due to paralysis of the dilator of the pupil from disease of the spinal cord. He had already had a case of spinal myosis under his care, and had seen several at Remak's clinique in Berlin. He was also acquainted, when he published his case, with Romberg's statement that in tabes there may be "a change in one or both pupils, consisting in contraction with loss of motion." The "Argyll-Robertson symptom," therefore, is the reflex iridoplegia, and is not necessarily associated with myosis, as might be inferred from some text-books, though, of course, myosis often is present.

"Gastric crises" were so named by the late M. Charcot, who states, in his *Lectures on Diseases of the Nervous System*,[27] that their connection with tabes was first pointed out by Delamarre in 1866. Charcot himself lectured on them in 1868. But Gull had suggested, as early as 1858 (as Charcot points out), that attacks of vomiting in a case of disease of the posterior columns were probably attributable to the spinal lesion. Gull made this suggestion in connection with his case of locomotor ataxy, to which reference has been already made. Topinard also, in 1864, remarked upon the occurrence of these attacks in tabes, but considered them to be merely a complication.[28]

Féréol described "laryngeal" or "bronchial crises" in 1868, in a collective work on the subject of these complications of tabes. He recalled the fact that Cruveilhier recorded a case of this kind in 1825.

In 1876, Raynaud and Lereboullet described "nephritic" or nephralgic crises. Attacks of pain in the face and head were described by Pierret in 1876; but severe pain in the head was a symptom in Duchenne's first case.

In 1879, Berger and Rosenbach, of Breslau, called attention to the relative frequency of aortic insufficiency in cases of tabes.[29]

A great variety of trophic lesions are now recognised as occurring in cases of locomotor ataxy.

Tabetic arthropathy was first described by Charcot in 1868.[30] Weir Mitchell had recorded, in 1831, two cases of

Pott's disease complicated with arthropathy;[31] and arthropathies of the *paralysed* limbs in paraplegia had been described by Scott Alison in 1846.[32] Suppuration of a tabetic joint is exceptional, but such an occurrence was described by Bourceret in 1875.[33]

Spontaneous fractures of bone in tabid patients were described in 1873 by Charcot in France, and by Weir Mitchell in America, almost simultaneously. A case has been already quoted from Cruveilhier, who wrote at a much earlier date, but who was not then in a position to appreciate the significance of the symptom, though he described it faithfully.

The "tabetic foot," or "tabetic club-foot" ("pied tabétique"), due to the occurrence of changes in the tarso-metatarsal articulations, was so designated, in 1883, by Charcot and Féré, when describing the four cases of this lesion known at the time.[34]

For our knowledge of the pathology of perforating ulcer of the foot, a well-known complication of tabes, we are indebted to Duplay and Morat.[35] Nelaton had described a case in 1852. Immediately afterwards, Vesignié recorded other instances (1852), and gave to the condition the name "mal plantaire perforant." He considered it to be a variety of psoriasis. Later writers, *e. g.*, Pean (1863) and Raynaud (1865), thought it was due to atheroma of vessels. Lucain, whilst recognising this as the usual cause, suggested that a few cases might be due to nerve degeneration. Duplay and Morat (1873) showed that changes in nerves or in the central nervous organs are the cause of all cases. Among the numerous examples collected by them, several were in subjects of tabes. Paul Bruns, of Tübingen, published important papers[36] on this subject in 1875. While the value of Bruns's contribution is recognised in France, this author, on his part, states that the disease is a French discovery.

Desprès has recorded a case of perforating ulcer of the hand; this is a rare thing. In the same communication, he records a case of perforating ulcer of the foot in a woman—an occurrence which he styles "une rareté parmi les raretés."[37]

I may here allude to a rare case which I saw at Professor Fournier's clinique in Paris, in 1889. It was one of symmetrical perforating ulcers, one of which was situated on the inner aspect of each buttock.

In 1881, Strauss described "tabetic ecchymoses" ("ecchymoses tabétiques"), which may occur in any part of the body affected by lightning pains.[38]

Charcot pointed out[39] that muscular atrophy is not uncommon in tabes, and he referred to a communication by Pierret in 1870. Leyden (1883) called attention to the changes which sometimes occur in the cells of the anterior horns of the spinal cord. Westphal first (1878), and afterwards Pierret (1880), described changes in the cutaneous nerves in cases of this disease. Déjérine (1883) did much to show the importance of these peripheral neurites in tabes, and remarked on the part which they probably play in the production of inco-ordination. The work of Pitres and Vaillard (1883) ought also to be mentioned here. In a later study[40] (1884), Déjérine proposed the name "peripheral neuro-tabes" ("nervo-tabes périphérique") for cases of peripheral neuritis with symptoms resembling those of locomotor ataxy. Still later (1889), the same author described[41] muscular atrophy as occurring in tabes in consequence of disease of peripheral motor nerve-fibres.

The somewhat equivocal position still occupied by deafness as a symptom of tabes, makes it of minor importance for us at present. But it may be stated that, in 1882, Strümpell published a case of complete deafness from atrophy of the acoustic nerves. In 1866, Lucae had mentioned two cases of deafness in tabes, and, in the first case described by Duchenne, hearing was quite lost in one ear.

While, in typical cases of locomotor ataxy, the mental faculties remain unaffected, it has long been known that this is not always so. According to Raymond, a case was recorded by Horn in 1833, in which dementia paralytica was associated with locomotor ataxy. Since then, many alienists, particularly in France, have published cases in which delirium, hallucinations, &c., have occurred. Hoffmann, Türck, Baillarger (1862),

Magnan, Falret, Topinard (1864), Jaccoud, Luys, and Foville fils (1873) are some who may be named. We are indebted to Westphal, perhaps more than to any other recent writer, for our knowledge of this subject.

In 1881, Westphal published a case which is of special interest, on account of the opportunity it furnished him of investigating the very earliest lesions of tabes. The patient was the subject of dementia with délire des grandeurs, and amaurosis. He was watched during two years for the first indications of tabes. At length one knee-jerk was lost, and soon afterwards the other. The patient died within a few weeks. The spinal cord was normal to the naked eye, but after hardening, some degeneration was found in the lateral columns and in Burdach's column.

Rokitansky and Türck (in the sixth decade of this century) are to be credited with taking the lead in studying the histological characters of posterior sclerosis. This they did in well-marked and advanced cases of the disease; but modern diagnostic methods have given us the opportunity of recognising the disease in its very early stage, and so of examining, on rare occasions, a cord which has as yet undergone but little change. The early lesions were investigated in this way, about 1872 and 1873, by Pierret, who worked under the direction of Charcot.[42] Pierret concluded that the disease begins with two symmetrical islets of sclerosis in the outer divisions of the posterior columns. (The "postero-external columns" are also called "Burdach's columns," or the "posterior root-zones;" or, in French, "bandelettes externes," "rubans externes," or "zones radiculaires postérieures.") He said that this was the essential lesion, and that the sclerosis of Goll's columns was a secondary degeneration. Charcot and Vulpian endorsed Pierret's view; but Strümpell, of Leipsic, showed that the innermost part of Goll's column is, in some cases at least, sclerosed at the very outset.

The discovery by Lissauer of the tract of fibres known by his name may be mentioned here, as it was first described (in 1885) in connection with its disease in tabes.[43]

Writing in 1863, Leyden stated that the difference between

inflammation and sclerosis of the spinal cord was so marked, as to justify us in separating diseases of the cord, as Vulpian had proposed, into two great classes—viz., funicular or fascicular (strangförmig) and focal (herdförmig). The latter are more inflammatory, the former more degenerative in their nature. The funicular differ from the focal group in involving a much greater length of cord, and in affecting particular fibre-tracts. As Flechsig named sets of fibres which are alike in course and function, "fibre-systems," so Leyden proposed to name diseases of these fibre-systems, "system-diseases" of the cord.[44]

The difficulty experienced by an ataxic patient in walking backwards ("Althaus's symptom") was pointed out by Dr. Althaus in 1884.[45]

One of the most important recent controversies in connection with tabes has been with regard to the influence of syphilis as a predisposing cause. Every one admits that the tabes of a syphilitic is the same as that of a non-syphilitic person, and that there is nothing specifically syphilitic in the morbid anatomy of locomotor ataxy. Duchenne recognised, in 1859, that syphilis sometimes precedes ataxy, and also that anti-syphilitic treatment does not cure tabes. In 1871, Dr. Buzzard included progressive locomotor ataxy among the nervous affections belonging to the tertiary stage of syphilis.

The fact of the association of tabes with late syphilis, noted as it had been by Duchenne, Lancereaux,* and Schultze (1867), was brought prominently forward by Fournier in 1875. In 1878, Dr. Gowers asserted, at the meeting of the British Medical Association, that syphilis is the cause of half the cases of locomotor ataxia. This view was supported by Vulpian in 1879, and in the same year by Erb, who, when he wrote the article in Ziemssen's *Cyclopædia* (1876), had been in doubt on the subject.

* Lancereaux, however (though he may have admitted the *association* occasionally as a fact), was strongly and emphatically opposed to the idea of a syphilitic *causal* relation, as you will find on consulting his works, and also (I think) in the discussion of Erb's paper at the London International Congress, 1881, at which Lancereaux was present.—W. T. G.

The opposite side was taken by Westphal, Remak, and Bernhardt (all in 1880).

Dr. Buzzard (1882), adding together the statistics of Fournier, Erb, and himself, obtains a syphilitic percentage of 59·8 in tabes; yet he considers the causal relationship between the two to be doubtful.[46]

Hippocrates, or rather the pseudo-Hippocratic writers, attributed tabes dorsalis to sexual excesses,* and it was only in the present century that a direct and close relationship between the two was disproved. But within recent years syphilis has come in as one of the most important etiological factors; and as syphilis is, in men—and it is they who are the principal sufferers from tabes—almost invariably due to deviation from strict sexual morality, it may be that, after all, the writers of the pre-Christian era were not entirely in error. Thus, increase of our knowledge brings us back to where we were thousands of years ago; for, as Sir Thomas Browne remarks,[47] "old Truths voted down begin to resume their places." Yet we do not return exactly to the same position. As the old man Clifford (in *The House of the Seven Gables*) puts it, in one of his lucid intervals, "All human progress is in a circle; or, to use a more accurate and beautiful figure, in an ascending spiral curve. While we fancy ourselves going straight forward, and attaining at every step an entirely new position of affairs, we do actually return to something long ago tried and abandoned." The same idea has been expressed by different writers when dealing with very different subjects.

* I wish you had attempted to show from actual quotations what amount of real substance there was in those old post-Hippocratic views of the *φθίσις νωτίας*.

My own impression is that there was very little, and that, in clinging to the mere *name* of tabes, or tabes dorsualis, the Germans have simply perpetuated superstition and confusion.

This is the more remarkable, as Romberg, while using, or indeed reviving the name, emphatically protested against the theory of sexual abuse and emissions which it implied.—W. T. G.

Locomotor ataxy possesses a melancholy interest for those of our profession who are admirers of Heinrich Heine. The great German poet, after terrible suffering, died of this disease in Paris, on the 17th February, 1856.[48]

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[3] Dechambre's *Dictionnaire encyclopédique des Sciences Médicales*, 1885, sér. iii, tom. xv.

[4] Hippocrates, *Of Diseases*, Bk. II; *Of Internal Affections; Of the Places in Man.* It is doubtful if any one of these writings is by Hippocrates himself.

[5] The story of Democrates, with other interesting information, is given by Neale in the work referred to. A third edition was published in 1806.

[6] Translated by Sydenham Society, 1853; see vol. ii, p. 395 *et seq.*

[7] Article "Tabes dorsalis," in Eulenburg's *Real-Encyclopädie*, Bd. xiii.

[8] Ziemssen's *Cyclopædia*, English translation, 1878.

[9] J. Bouillaud, *Traité de Nosographie Médicale* (Paris, 1846), tom. v, p. 317 *et seq.*

[10] Ph. Hutin's "Investigations and Observations on the Pathology of the Spinal Cord" were translated into German for Nasse's *Sammlung* (1838). The observations seem to have been made in the years 1821 to 1825. They appeared in the original French about 1828, but I have not had access to them in this form.

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[12] *Medico-Chirurgical Transactions*, 1840, vol. xxiii, p. 80; *London Medical Gazette*, 1840, vol. xxv, p. 763.

[13] *Cyclopædia of Anatomy and Physiology*, 1847, vol. iii, pp. 721 O—R.

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[15] Romberg, *op. cit.*

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[18] Duchenne; see *Selections from the Clinical Works of Duchenne*, New Sydenham Society, 1883, p. 33.

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[21] *Selections from the Works of Duchenne*, New Sydenham Society, 1883, p. xvii.

[22] *Ibid.*, p. 35.

[23] "Ueber ein frühes Symptom der Tabes dorsalis," *Berliner Klin. Woch.*, 1878, No. 1.

[24] *Berliner Klin. Woch.*, January, 1881, Nr. 1-2.

[25] *Deut. Archiv f. Klin. Med.*, 1883, Bd. xxxiii, S. 192.

[26] *Edinburgh Medical Journal*, February, 1869.

[27] Translated by New Sydenham Society, 1877.

[28] Gowers, *Diseases of the Nervous System*, second edition, 1892, vol. i, p. 413, note.

[29] *Berliner Klin. Woch.*, 1879, S. 402.

[30] *Lectures on Diseases of the Nervous System*, New Sydenham Society, 1877.

[31] See Raymond's article in Dechambre's *Dictionnaire* (*vide supra*, [3]).

[32] Charcot, *loc. cit.*

[33] *Progrès Médical*, No. 41, October, 1875, p. 573 *et seq.*

[34] Féré, *Rev. de Méd.*, 1884, tom. iv, p. 473; *Brain*, January, 1885, vol. vii, p. 546.

[35] *Arch. gén. de Méd.*, 1873, tom. i.

[36] *Berliner Klin. Woch.*, 1875, Nr. 30-32.

[37] *Gaz. des Hôp.*, 1880, p. 1019.

[38] See *Brain*, January, 1885, vol. vii, pp. 544-549.

[39] *Lectures on Diseases of the Nervous System*, second series, New Sydenham Society, 1881.

[40] *Arch. de Physiol.*, 1884, tom. i, pp. 231-268.

[41] *Rev. de Méd.*, 1889, p. 81 *et seq.*

[42] Charcot, *Lectures on Diseases of the Nervous System*, second series, New Sydenham Society, 1881, p. 10.

[43] Gowers, *op. cit.*, vol. i, pp. 421, 422, note.

[44] Leyden, *loc. cit.*

[45] *British Medical Journal*, October, 1884, p. 708; *On Sclerosis of the Spinal Cord*, 1885, p. 227.

[46] For connection between syphilis and tabes, see Gowers, *Lancet*, January, 1881; *Diseases of the Nervous System*, vol. i, p. 426; *Syphilis of the Nervous System;* Buzzard, *Clinical Lectures on Diseases of the Nervous System*, 1882.

[47] *Christian Morals*, pt. ii, sec. v.

[48] See Althaus, *On Sclerosis of the Spinal Cord*, p. 273.

II.

HISTORY OF PRIMARY SPASTIC PARALYSIS.

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HISTORY OF PRIMARY SPASTIC PARALYSIS.

NOMENCLATURE.

Primary spastic paraplegia.
Primary lateral sclerosis.
Primary and symmetrical sclerosis of the lateral columns of the spinal cord (*Charcot*).
Spasmodic tabes dorsalis (*Charcot*, in contrast to ataxic tabes dorsalis).
Spastic spinal paralysis (*Erb*).
Erb's palsy.

HISTORY.

FIFTY years ago, Abercrombie described a typical case of spastic paralysis, with involvement ultimately of the arms as well as the legs. The patient died of phthisis two years after the commencement of the disease, and no changes were found in the brain or spinal cord.[1]

If other cases of this kind were published, they, as well as this one, were overlooked by subsequent observers. For, imperfect as our knowledge still is of the anatomy of spastic paralysis, it is only twenty years since it first took a definite place in our Nosology as a clinical entity.

Speaking of chronic spinal meningitis, Dr. Gowers remarks[2] that formerly "it was inferred that chronic symptoms analogous to those which, when acute, are due to acute meningitis, were evidence of a chronic form. This was, therefore, assumed to be the cause of a group of symptoms, of which spasm is the prominent and dominant feature, which are now known to be due solely to a morbid state of the spinal cord itself, and to be consistent with a perfectly normal state of the

membranes. Hence 'chronic meningitis,' as it was recognised twenty years ago, as a 'clinical entity,' has ceased to exist, or rather has passed into the pathological conception of 'primary lateral sclerosis.'"

There is little doubt, however, that Erb disentangled spastic paraplegia from a mass of clinical types which included many diseases besides chronic meningitis.

The occurrence of secondary degeneration of nerve-tracts after a lesion of the central nervous system was first shown by Rokitansky in 1847; but we are indebted still more to a later observer, Türck, for the anatomical knowledge that has been gained in this way. The latter (1856), after investigating the nature of secondary degeneration, published several cases in which degeneration of the pyramidal tracts was discovered without any gross lesion to account for it. Charcot published, in 1865, a case in which the lateral columns only were sclerosed. The grey matter and its cells were intact, but several of the anterior roots were atrophied.[3]

Erb was the first (1875) to separate spastic paraplegia as an individual type from the midst of a heterogeneous group of chronic nervous diseases.[4] His communication was based on about a dozen cases without any autopsy; in only two cases were the arms affected. The symptoms described included exaggeration of the tendon-reflexes. (The tendon-reflexes were first investigated by Erb and Westphal shortly before this.) In the light of Charcot's researches of 1865—which, however, Erb thought must have been carried out on old and complicated cases—the latter considered it probable that the lesion in his own cases was a primary sclerosis of the lateral columns.

In a lecture delivered in 1876, Charcot gave a full account of the disease, which he called "spasmodic tabes dorsalis," by way of contrast to what he termed "ataxic tabes dorsalis" (locomotor ataxy). He fully admitted that Erb was the first to investigate the subject thoroughly.[5]

Charcot took up, on the whole, the same attitude as Erb, but was, if anything, less certain about the morbid anatomy. He said that the cases where the symptoms during life and

the appearances after death indicated lesion of the lateral columns, without participation of the anterior cornua, were all "old memories which have become indistinct, and require to be refreshed."

In the following year, Erb published another contribution [6] on the same subject, and speaking of the time before the date of his former article, used these words—"Von der wirklichen, primären, lateralen Sclerose, existirte in der Literatur nicht ein einziges Beispiel." Some sentences later, however, he qualified his words a little by adding, "am wenigsten bei uns in Deutschland." I have already pointed out that Abercrombie recorded the clinical features of a case as early as 1845, thirty years before the time Erb refers to. As, however, this author, though he suspected the existence of disease in the spinal cord, could find after death no trace of changes in either brain or spinal cord, this observation of his detracts in no way from the credit due to Erb as having been the first to indicate the actual seat of lesion.

Our knowledge of the morbid anatomy of Erb's paralysis is still scanty indeed. It so happens that in almost every autopsy on a case which, during life, has presented the symptoms of this malady, the changes have been found not to be confined to the pyramidal tracts. One reason of this defect in our knowledge is that the disease has so little tendency to shorten life; it is not associated with those secondary disorders of the urinary, respiratory, and other systems which often cause death in tabes, progressive muscular atrophy, &c.; and another reason is that the degeneration seems to begin at the lower end of the pyramidal tracts—viz., where the fibres divide and form delicate arborisations and plexuses in the grey matter around the motor cells of the anterior horns; this is, of course, the region where the fibres are furthest removed from their nutrient cells in the cerebral cortex.

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III.

HISTORY OF ATAXIC PARAPLEGIA.

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HISTORY OF ATAXIC PARAPLEGIA.

NOMENCLATURE.

Combined lateral and posterior sclerosis.
Combined postero-lateral sclerosis.
Ataxo-paraplegic tabes (*Déjérine*).
Ataxic paraplegia (*Gowers*).
Ataxo-spastic tabes (*Grasset*).
Combined fascicular sclerosis.
Progressive spastic ataxia (*Dana*).
Postero-lateral spinal (or fascicular) sclerosis (*Damaschino*).

HISTORY.

Writers have hitherto generally traced back the history of this disease to the works of Prévost and others, in 1877, and of Westphal, in 1878. It ought to be mentioned, however, that Westphal described its occurrence in the insane at a much earlier date—viz., in 1867. Having already published a series of cases in which ordinary tabes dorsalis (posterior sclerosis) was associated with mental disease, he endeavoured, in an article, "Ueber Erkrankungen des Rückenmarks bei der Allgemeinen Progressiven Paralyse der Irren," to demonstrate that disturbances of motility in insane patients are due, not to brain disease, as had heretofore been supposed, but principally to disease of the spinal cord, which may sometimes extend up to the medulla and pons. Westphal's cases of spinal lesion in general paralytics included several examples of sclerosis of the lateral columns only, and three instances of combined sclerosis of the posterior and lateral columns. With regard to these latter, Westphal remarked

that the gait was not such as is usual in posterior sclerosis—that the patients did not reel when they shut their eyes; but it must be borne in mind (he said) that the affection of the posterior columns was in the upper part of the spinal cord, and was slight in two of the cases.[1]

When Westphal was studying the knee-jerk, in 1875, he found it to be absent in tabes, and also in combined sclerosis, if the posterior sclerosis involved the lumbar region of the cord.[2]

Apart, however, from general paralysis of the insane, the principal early writers on combined posterior and lateral sclerosis were Prévost (1877), Pierret, Kahler, and Pick (1878), Westphal (1878), and Babesiu of Buda-Pesth (1879). Babesiu's case was one of sclerosis of the lateral columns, and of Goll's columns, with optic atrophy.[3]

An interesting article, from the pen of Damaschino, appeared in 1883. In addition to various other combined scleroses, he described cases of: (1) general paralysis with lateral sclerosis; (2) general paralysis with posterior sclerosis, lateral sclerosis, and optic atrophy; and (3) posterior sclerosis with lateral sclerosis. Damaschino said that this last combination was undoubtedly of more frequent occurrence than might be inferred from published accounts of it.[4]

A case of some importance was recorded by Ballet and Marie in 1884. There was sclerosis of the posterior and lateral columns with optic atrophy.[5]

Ataxic paraplegia owes its establishment as a definite morbid type, on both anatomical and clinical grounds, to the complete picture of it drawn by Déjérine in 1884 under the name of "ataxo-paraplegic tabes." Independent descriptions were given, two years later (1886), by Gowers and Grasset, each of whom proposed a new designation for this affection.[6]

Déjérine thought—at least when he first wrote on this subject—that the sclerosis began in the posterior columns, and spread to the lateral columns by way of the meninges. Westphal seems to have considered this possibility at an earlier date, and to have put it aside on the ground that the meningeal inflammation did not correspond, in situation and

intensity, with the lateral sclerosis. He took the lateral sclerosis to be the primary lesion.[7]

An important paper on this subject was written by Dana, of New York, before Gowers's lecture on ataxic paraplegia appeared in print, but was not published till the following year (1887). Dana arranged his cases of what he termed "progressive spastic ataxia" into two groups: (1) those in which motor or spastic symptoms predominated, and (2) those in which sensory or tabid symptoms were more conspicuous. He thought that the typical lesion in this disease was a sclerosis of three long fibre-systems of the cord:—the columns of Goll, the direct cerebellar tracts, and the pyramidal tracts. A suitable designation, therefore, for the disease would be "combined fascicular sclerosis." A view similar to this appeared to commend itself to Strümpell.

Attention may be called to a later paper (1889) by Dana, who has by this time experience of fifteen cases. Though here (in a British journal) adopting the name proposed by Gowers, he is still inclined to think that the designation he originally employed is the more accurate one, because, in his experience, spastic ataxia is the leading symptom, whilst paralysis is but slight, in cases of combined posterior and lateral sclerosis.[8]

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[5] See *Brain*, January, 1885, vol. vii, p. 556.

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[7] See Ormerod, "On the Combination of Lateral and Posterior Sclerosis in the Spinal Cord," *Brain*, 1885-86, vol. viii, p. 110.

[8] Dana, "Progressive Spastic Ataxia (Combined Fascicular Sclerosis) and the Combined Scleroses of the Spinal Cord," *New York Medical Record*, 2nd July, 1887, vol. xxxii; *Brain*, January, 1889, vol. xi, p. 490.

IV.

HISTORY OF HEREDITARY ATAXY.

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HISTORY OF HEREDITARY ATAXY.

NOMENCLATURE.

Friedreich's disease (*Brousse*).
Hereditary ataxy (*Eisenmann*).
Postero-lateral spinal sclerosis of generic origin (*Everett Smith*).
Generic ataxy (*Everett Smith*).

HISTORY.

This disease was first separated from ordinary tabes by Friedreich, of Heidelberg, in an address delivered to the Congress of German Naturalists and Physicians in September, 1861. This communication was not published till 1863.[1] Meanwhile a remarkable case had been recorded by Marius Carré in a thesis published in Paris in 1862.

Friedreich's description of the disease was fairly complete. He noted the commencement with weakness of the lower limbs; the muscular inco-ordination, which was even greater than the want of power; the disturbance of speech; the spinal curvature, nystagmus, and giddiness; and the frequent commencement at puberty or in early adolescence. He also observed the deformity of the foot, and insisted on the etiological importance of alcoholism in the parents. He recognised the affection clinically as a chronic progressive loss of motor co-ordination; and anatomically as a chronic degenerative atrophy, mainly of the posterior columns, but also, to some extent, of the lateral columns, beginning in the lumbar region, and ceasing after reaching the medulla, and involving the nucleus and fibres of the hypoglossal nerve.

The disease, then, was at first regarded as a particular variety of the progressive locomotor ataxia of Duchenne. Friedreich said, in the account of his first published cases, that he considered himself entitled to separate this well-marked group (which consisted of two members of one family and four of another) from the general designation "tabes dorsalis." Some writers thought, after Vulpian and Charcot had described multiple sclerosis, that the hereditary form of ataxy was a variety of disseminated sclerosis, or a combination of this disease with locomotor ataxy.

Friedreich published two further contributions to this subject in 1876 and 1877.[2] In them he continued the histories of those of his patients who still survived at the date of his former article, and he also gave new cases. Of the total of nine cases, six suffered and five died from typhus abdominalis; and he therefore judged that ataxy must diminish the power of resistance to the onslaught of this disease. He noted how different, with regard to inherited predisposition, sex, and age, the hereditary was from the more usual form of ataxy. He found the tendon-reflexes abolished in the four cases examined. (These reflexes, which were only described by Erb and Westphal in 1875, were, of course, not known when Friedreich wrote on the former occasion.) One of Friedreich's autopsies was on a case of twenty-three years' standing.[3] Grey degeneration was found in the posterior columns and in the hinder part of the lateral columns. The spinal ganglia, peripheral nerves, and muscles of the extremities were normal. The medulla oblongata, especially in its lower part, was distinctly diminished in volume. It seems most probable, the author says, that complete formation and development of the cord and medulla did not take place, and that a sclerosis occurred in these defective structures. This sclerosis must have set in not later than the time when the first distinct morbid symptoms began.

The first time the disease was recognised and described in England was in 1880. In October of that year, before the Clinical Society of London, Dr. Gowers "brought forward

five cases of locomotor ataxy in members of the same family, three of whom were exhibited."[4] Dr. Gowers alluded to the fact that, in 1871, Dr. A. Carpenter, of Croydon, showed two sisters, who were evidently suffering from this complaint, to the Medical Society of London. A brother in this family had since then become affected.

In 1882 an important study of Friedreich's disease was published by Dr. Auguste Brousse, who first named it after its discoverer. Brousse pointed out that the two sexes appear to be equally predisposed, and he distinguished the affection from tabes, from multiple sclerosis, and from the combined sclerosis of the columns of the cord which had been described by Prévost, Westphal, and others.

In 1883, Rütimeyer, of Basel, published a series of cases which were possessed of features so curious and interesting that they may be mentioned here. The eight patients belonged to four families named Blattner, and were all directly descended from one Blattner, who married in 1710. This common ancestor was known as Blattner Stülzi, "Stülzi" being equivalent in the vernacular of his district to "Stolperer" (English, "stumbler") in a less Doric dialect of German. Those of his direct descendants from whom the four families were derived were brothers about the beginning of this century. To distinguish this line of Blattners from other Blattners of the same village of Küttigen, all its members were called "Stülzi," not only in popular talk, but also in family Bibles and in official church registers. This continued till the fifth decade of the present century, so strongly had the stumbling gait of the original Blattner Stülzi impressed itself upon the memories of his contemporaries in the district where he lived.

Rütimeyer's eight patients were all born within the twenty years 1848 to 1868, and the youngest began to suffer twenty years after the oldest.[5]

On the occasion of a clinical lecture at the Salpêtrière in 1884, Charcot showed a patient with hereditary ataxy, and expressed the view that this disease is different from both locomotor ataxy and disseminated sclerosis. Till then,

apparently, he had never committed himself to an opinion on the nature of the cases described by Friedreich.

Reference may be made to a paper by Dr. Ormerod, "On the so-called Hereditary Ataxia, first described by Friedreich" (1884).[6] Here may be found tables of the genealogical relations of about forty cases, with accounts of the individual cases in abstract, and the literature to date.

Another important account of Friedreich's disease is due to W. Everett Smith in America. He reported six cases in girls of one family, all the boys remaining unaffected. The first symptoms in each instance were: gastric disturbance and palpitation, followed by unsteady gait; ataxy gradually extending to the hands, the disease thereafter remaining stationary for an indefinite period. There was a well-established history of alcoholism in previous generations. One of the patients only began to suffer at the age of sixty-six.

At the meeting of the Society to which Everett Smith read his paper (9th June, 1885), Dr. Morton Prince reported a sporadic case of this disease. The ages, however, of the other children of the family were such that they might possibly yet suffer.[7]

In "Some Further Observations on Friedreich's Disease" (1888), Dr. Ormerod described some cases, and emphasised the fact that, while family proclivity is a distinguishing feature of the disease—the one that caused Friedreich to separate it from tabes—sporadic cases do nevertheless occur; and that, moreover, the symptoms are now so well known that such cases can be diagnosed. Dr. Ormerod also emphasised the etiological importance of the acute specific fevers, and of intemperance in the parents.[8]

Dr. Ladame's address to the Medical Society of Geneva[9] is a contribution of great value. While not, perhaps, containing much that is original, it is an admirable critical review of our present knowledge of the subject; and the bibliography which is appended (embracing over a hundred references) ought to be useful to those who are interested. Up till this date we have records of nine autopsies on authentic cases.

Finally, in September, 1893, Dr. J. Wallace Anderson described two cases (brother and sister) which, he said, were the first recorded in Scotland.[10]

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[6] *Brain*, April, 1884, vol. vii.

[7] The cases of Everett Smith and Morton Prince were published in the *Boston Medical and Surgical Journal*, October, 1885, vol. cxiii, pp. 361, 371.

[8] *Brain*, January, 1888, vol. x.

[9] Translated in *Brain*, 1890, vol. xiii.

[10] *Glasgow Medical Journal*, 1893, vol. ii, p. 168.

V.

HISTORY OF PROGRESSIVE MUSCULAR ATROPHY.

V.

HISTORY OF PROGRESSIVE MUSCULAR ATROPHY.

NOMENCLATURE.

Progressive muscular atrophy (*Aran*).
Atrophic muscular paralysis (name given by Thouvenet and adopted by Cruveilhier).
Chronic spinal muscular atrophy.
Chronic protopathic spinal amyotrophy (*Charcot*).
Chronic anterior poliomyelitis (*Charcot*).
Chronic parenchymatous tephromyelitis (*Charcot*).
Progressive muscular atrophy of the Duchenne-Aran type (*Charcot*).
N.B.—Charcot recognises as distinct from this the disease which he names "amyotrophic lateral sclerosis;" he places the latter in a group to which he gives the designation "deuteropathic spinal amyotrophies."
Progressive fatty muscular atrophy (*Duchenne*).
Cruveilhier's disease or atrophy.
Creeping, or partial, or local, or wasting palsy.
Chronic progressive polymyositis (*Friedreich*).

HISTORY.

In 1830, Sir Charles Bell recorded[1] a few cases of paralysis of the muscles of the extremities. It is very doubtful if any of them can be classed with the disease whose history is now being traced. One of them, at least, is undoubtedly an instance of primary myopathy.

In the following year, Darwall of Birmingham published[2] a series of cases, of which two, at least, appear to have been genuine examples of progressive muscular atrophy. Indeed, Darwall's description is so intelligible to a modern reader

that it would seem quite fair to attach his name to the disease, instead of that of Cruveilhier, who wrote about it at a date so much later.

In 1844,[3] Bell published several new cases of muscular atrophy. Probably a few are instances of progressive muscular atrophy. He supposes these affections of the muscular system of the extremities to be connected in some way with the sympathetic system of nerves, and, through that system, with the bowels. This allusion to the sympathetic foreshadows the promulgation of a theory which is commonly supposed to be of much more recent origin.

Graves records[4] a case of muscular atrophy involving the left upper arm and the right fore-arm. Some uncertainty is, perhaps, attached to this case, owing to the fact that the disease was arrested, though not cured, by mercurial treatment.

Judging from some of the names that have been bestowed upon the disease, it might be inferred that the history of progressive muscular atrophy only began after the time of Graves's work. Aran's well-known paper, entitled "Recherches sur une Maladie non encore décrite du Système Musculaire (Atrophie Musculaire Progressive)," appeared in September and October, 1850.[5] Some details about this communication are of importance. Aran states that he had published, in *L'Union Médicale*, nearly two years before, the first case of the kind that had come under his notice. He acknowledges his immense indebtedness to his friend Duchenne for placing at his disposal all the facts that he (Duchenne) had collected, and for assistance so great, in investigating the muscular systems of his patients, that he simply could not have done without it. Aran acknowledges his indebtedness to the writings of Van Swieten, Graves (1848), Darwall (1831), Dubois (1847), but, above all, to the writings of Charles Bell and Abercrombie (1845). He brings together in his paper eleven cases, of which, however, we must, with our present knowledge, reject several; for cases that occur in early life, in several members of a family, and without anatomical changes in the nerves connected with the affected muscles, are now recognised as belonging to a different category.

Aran gives a good description of progressive muscular atrophy, and concludes that it may occur apart from any lesion of the central or peripheral nervous system, and without any arrest of the circulation.

The next contribution that must be mentioned is one by Cruveilhier, "Sur la Paralysie Musculaire Progressive Atrophique."[6] As early as 1832, Cruveilhier had seen a lady who suffered from what we can now easily recognise as a combination of progressive muscular atrophy and bulbar paralysis. This patient died about a year afterwards, and Cruveilhier was astonished to find, at the *post-mortem*, no lesion of the central nervous system. Never before had he been so disheartened at the apparent helplessness of pathological anatomy in the case of certain affections of the nervous system. For a long time—viz., while he was associated with the Salpêtrière—he saw no other similar case; but after he became connected with the Charité, he saw several. The first that came to *post-mortem* was that of a lad aged eighteen.[7] Cruveilhier found the brain and cord healthy, the nerves as bulky as normally, and the muscles atrophied (April, 1848). He concluded that the cause of the palsy was in the muscles themselves; the muscles wasted, and the degree of weakness was exactly proportional to the amount of wasting.

Cruveilhier states that he took every opportunity of making known the results of these two autopsies. Thus, in his summer course of 1848, he lectured on "L'Atrophie Musculaire Primitive ou Idiopathique" as the cause of a certain variety of paralysis.

In the beginning of 1849, Duchenne communicated to the Institute a memoir on "Atrophie Musculaire avec Transformation Graisseuse," in which he said he was indebted to Cruveilhier for his knowledge of the morbid anatomy of this disease. Duchenne communicated to Aran the results of Cruveilhier's second autopsy; and, in September, 1850, Aran published his paper on "Atrophie Musculaire Progressive," with a collection of cases. On the recommendation of Cruveilhier, Thouvenet, his interne, took up the subject for

his Inaugural Thesis, which was produced in 1851, under the title of "Paralysie Musculaire Atrophique."*

From a third *post-mortem*, which he made in February, 1853, Cruveilhier inferred that the primary lesion of progressive muscular atrophy was not atrophy of the muscles, but atrophy of the anterior roots of the spinal nerves. The case was one of combined spinal and bulbar muscular atrophy. The orbicularis oris, particularly in the lower lip, was atrophied, and the tongue had lost much of its muscular tissue. In connection with this, it was observed that the hypoglossal nerve was so wasted as to present a striking contrast to another nerve which supplies this organ—the lingual, which was well preserved. Cruveilhier put the pathological sequence thus:—diminution of nervous impulses, owing to atrophy of anterior roots; diminished contractility; muscular atrophy. Atrophy of the anterior roots, he thought, was independent of any appreciable lesion of the cord.

In 1853, Duchenne published a paper,[8] which is of interest (apart from the records of cases) because in it he tries to show the great value of localised electrisation in the treatment of this affection. He also discusses here the question how, in view of Cruveilhier's discovery, faradisation of the atrophied muscles can arrest, or even remove, the wasting due to atrophy of the anterior roots, and why this treatment succeeds in some cases and not in others.

It took fifteen years to get one step further on, but the position that was thereby gained was of the first importance. What was arrived at was the theory, which has been but confirmed by the searching criticism of almost a generation, as to the morbid anatomy and physiology of the central nervous system in relation to this disease. The first discovery of changes in the cells of the anterior cornua of the spinal cord has been credited in England to Lockhart Clarke,[9] and

* There need be no dispute as to the fact of Duchenne laying before Aran the results of the autopsy made by Cruveilhier, for it is stated more or less directly by all the parties concerned. To judge from their writings, they had no dispute among themselves, though this seems to be hinted at in an English publication.

in other countries to Luys.[10] It is not absolutely certain that the discovery—so far as genuine progressive muscular atrophy is concerned—ought to be credited to either of these observers; but if one of them does deserve the honour, that one, it appears to me, is undoubtedly Luys.

Luys's paper, which appeared in 1860, was entitled "Atrophie musculaire progressive. Lésions histologiques de la Substance Grise de la Moelle Epinière."[11] The case was one of muscular atrophy in the left arm, the right being scarcely affected. The patient died of intercurrent pneumonia, and changes were found in the grey matter of the cord at the part corresponding to the nerve-supply of the affected muscles—viz., disappearance of many cells of the left anterior cornu, degeneration of neighbouring cells, and atrophy of the anterior roots. The details given of the symptoms were so scanty that Duchenne hesitated to admit the case as one of progressive muscular atrophy. Still, for the same reason, we are scarcely in a position to reject it.

On the other hand, the claim made on behalf of Lockhart Clarke is based on his reports on the morbid anatomy of three cases. The first (1861) was a very remarkable case, whose clinical features were recorded by Dr. Gairdner.* With

* It may be of some interest to you (Dr. Monro) to know what does *not* appear in print—viz., that my case (that of an old Scotch professor) was submitted in the course of one forenoon to *four* eminent London physicians; and the result of these four separate consultations and opinions was communicated to me verbally by the late Professor George Edward Day, of St. Andrews, who accompanied his colleague.

Of the four consulted (Copland, Burrows, Gull, and Todd), two thought it was lead-poisoning; two thought it was not.

Gull thought it was a spinal lesion in the region of the brachial plexus. Todd said, "If it's not lead, I don't know what it is." One of the others held out for lead so decidedly, that the poor professor was sent over to Aix-la-Chapelle to have it cleared out of him, and it was after all this that he came to me.—W. T. G.

Dr. Gairdner has recently informed me (verbally) that the doctors collected some gallons of the professor's urine and divided it into two parts. They sent one half to be examined at University College, and the other half to some other laboratory. The one analyst reported that the urine contained no lead; the other said it contained traces.—T. K. M.

regard to its genuineness, some doubt is introduced by the fact that severe pain was one of the most clamant symptoms. The changes observed after death included pigmentation of the cells of the anterior horns. The two other cases (1862, 1867) were not examples of progressive muscular atrophy. Duchenne and others have pointed this out, and probably no one will question the soundness of their judgment here. Of course the admission that these cases were not genuine does nothing to disprove the existence of a lesion in the anterior cornua in progressive muscular atrophy.[12]

Immediately after Clarke, mention ought to be made of Duménil, of Rouen, who recorded several examples of muscular atrophy, with damage to the cells of the anterior horns, and atrophy of the anterior roots. He added the weight of his influence to the side of the nervous as distinguished from the muscular theory of the disease. To explain the puzzling fact of the common absence of muscular wasting in the territory of the bulbar nerves in comparison with what occurs in other parts of the muscular system, he recalled an observation by Schneevogt, and suggested that the different relations between the sympathetic on the one hand, and the bulbar and the spinal nerves on the other, accounted for the discrepancy.[13]

Duménil attributed the progressive, fatty, muscular atrophy to nervous influence—namely, an influence emanating, not from the motor, but from other nerve-elements. He thought that one of his cases, in which the sympathetic nerve was profoundly altered (a case, however, which was not an example of progressive muscular atrophy), furnished conclusive evidence of the involvement of the sympathetic in this disease.

In a lecture delivered at the Salpêtrière, in June, 1868, Charcot expressed his belief that the so-called motor cells of the anterior cornua are the primary seat of mischief in progressive muscular atrophy and the spinal amyotrophies generally. Additional *post-mortem* observations and writings by Hayem (1869), Charcot and Joffroy (1869), and Duchenne and Joffroy (1870), established Charcot's doctrine on so firm a basis that it speedily found acceptance in the text-books.[14]

The idea that the great sympathetic nerve was one of the principal seats of lesion, if not the primary seat, has been variously attributed to Duménil (1867),[15] and, with more justice, to Schneevogt of the Hague (1854), who, in one instance, discovered an alteration of the splanchnic nerves. But I have already mentioned that Sir Charles Bell (1844) had recourse, at a much earlier date, to the same nerve for help in the solution of the same problem. Remak, Eulenburg and Guttmann were other advocates of the view that the sympathetic is responsible for the trophic disturbances. The writings of Charcot, Hayem, Duchenne and Joffroy (1869-1870), with the negative results—as regarded the gangliated cord—of a series of autopsies, appear to have been the agencies by which the death-blow was administered to the sympathetic theory.

Duchenne had taught, as early as 1849, that progressive muscular atrophy was, in the first instance, a peripheral disease. Aran, Virchow and Friedberg also considered the primary disease to be muscular. (But the case on which Virchow based his views is inadmissible, being a family case.)[16] Bell and Cruveilhier, on the other hand, believed that the nervous system was at fault. Unconvinced by Luys's observation (1860), Duchenne seems to have been first led to regard the disease as primarily spinal by Hayem's paper (1869); and, in the third edition of *L'Electrisation Localisée* (1872), we find him accepting the modern doctrine of the spinal origin of true progressive muscular atrophy.[17]

The current of opinion was now flowing strongly in the direction of the "nervous," and away from the "muscular" theory of the disease; yet the latter was not left without its supporters. One of the most determined of these was Friedreich of Heidelberg, who, in his large volume, *Ueber Progressive Muskelatrophie* (1873), stoutly opposed the view that the primary lesion was in the nervous system. He said the primary change occurred in the muscular tissue, the changes in the nerve-cells being secondary. In its pathological aspect, therefore, he considered the disease to be a chronic progressive polymyositis, and accordingly he would look upon

pseudo-hypertrophy as nothing other than a modified form of progressive muscular atrophy. But Friedreich's cases are rejected by Charcot[18] on the ground that the reports are not sufficiently complete; and even Friedreich's countryman, Erb,[19] remarks that his "weapons were not sharp enough; he based on a series of cases that was sadly in need of proper sifting, and contained a good deal that was foreign to the subject, and his microscopic work was not equal to the requirements even of that time."

Conversely, in his lecture on progressive bulbar paralysis, Kussmaul speaks of "pseudo-muscular hypertrophy" as of central origin, and proceeding from an atrophy of the anterior cornua and their ganglion-cells.[20]

A well-known case (which Erb subsequently identified as belonging to the "juvenile" group) was recorded by Lichtheim in 1878. Here there was no disease of the anterior cornua, and Lichtheim's investigation of the case made him a strong advocate of the myopathic theory of progressive muscular atrophy. He emphasised the close relationship between this disease and pseudo-hypertrophy, but yet believed that, on clinical grounds, they must be separated. Even Lichtheim's paper did not, immediately at least, alter the direction of the flowing stream.

Progressive muscular atrophy had hitherto been accepted as an unity in itself. But influences had already begun to work which were destined to bring about, before long, considerable modifications in professional opinion. In the effort to preserve this unity, writers had gone beyond the plain teaching of morbid anatomy. The unavoidable disintegration that followed resulted, however, not in chaos, but in the establishment, on a firmer basis than ever, of a scientific as well as practically convenient classification of the muscular atrophies. This disintegration was brought about by the detachment of one small group after another from the originally single, large, and heterogeneous group. It is not surprising that the first series of cases to be detached from the muscular atrophies were those of the pseudo-hypertrophic group. Duchenne had from the first (1861) recognised this to be distinct

from progressive muscular atrophy, though, as we have seen, Friedreich endeavoured, twelve years later, to unite the two forms again.*

In 1875, Leyden (then of Strasbourg) proposed to separate the "hereditary forms of progressive muscular atrophy" from those of the Aran-Duchenne type.[21] He pointed out the tendency of the hereditary form to commence in early life; to affect several members of a family, and especially those of the male sex; and to involve first the muscles of the lower limbs and sacral region. He also showed that the development of fat might mask a very considerable loss of volume of the damaged muscles. Leyden remarked further on the unmistakable resemblance between the hereditary and pseudo-hypertrophic forms. While suspending judgment on the question whether progressive muscular atrophy is primarily spinal or muscular, he remarked that all observations hitherto made seemed to indicate that pseudo-hypertrophy at least is of myopathic origin.

In 1879, Möbius[22] divided the chronic muscular atrophies into two groups: (1) hereditary—viz., pseudo-hypertrophy and Leyden's hereditary muscular atrophy; and (2) non-hereditary—viz., the Duchenne-Aran type. He said that though all our methods of examination may show that the brain and cord are quite normal in cases of the hereditary or degenerative type, we need, nevertheless, have no hesitation in looking upon this disease as a neuropathy, the primary flaw being central and not peripheral.

By this time, as Erb has remarked, neurologists had separated various secondary muscular atrophies from progressive muscular atrophy; and, in particular, those due to, or associated with, acute and chronic anterior poliomyelitis, multiple neuritis, syringomyelia, and articular affections. The originally single group, therefore, of the muscular atrophies

* Duchenne published the first description of pseudo-hypertrophic paralysis in the second edition of *L'Electrisation Localisée* (1861). He described it under the title of "Paraplégie hypertrophique de l'enfance de cause cérébrale." At a later date he substituted for this designation the name now commonly employed, "Paralysie musculaire hypertrophique."

had become greatly differentiated; and the series of cases to which the name of progressive muscular atrophy was still supposed to be applicable had become greatly curtailed.

In 1882, Erb detached from this attenuated group a third set of primary myopathies, which he termed the "juvenile form" of progressive muscular atrophy.[23] In cases of this type, the shoulder-girdle, upper arm, pelvic girdle, thigh and back suffer early, and the atrophy is often associated with true or false hypertrophy. Erb ranged, in one class, muscular atrophy of the undoubtedly spinal type (due to lesion of the anterior cornua); and, in another class, the hereditary, pseudo-hypertrophic and juvenile types. The question of the neuropathic or myopathic nature of the juvenile form must, he said, still remain quite open.

As early as 1855, Duchenne had shown that there is a particular variety of muscular atrophy which occurs in childhood. In 1872, in the third edition of *L'Electrisation Localisée*, he gave a new account of this "infantile" form, and pointed out that it begins in the face and is frequently inherited.[24] Erb did not deal with this form, but, in 1885, Landouzy and Déjérine published an important paper on it.[25] They included among its leading characteristics its commencement in infancy, its occurrence in families, and its tendency to involve first the face and subsequently the shoulder and arm. They therefore described it as the "facio-scapulo-humeral" type of "pure progressive atrophic myopathy." They mentioned the "facies myopathique" as one of its peculiarities, and they ranged the disease alongside the other three that had already been separated from spinal muscular atrophy.

Reference may be here made to one of the most important recent writings on the subject—Erb's lecture on "Progressive Muscular Dystrophy."[26]

Even an historical sketch would be incomplete if no allusion were made to a question which is still a matter of much controversy amongst neurologists—the identity or otherwise of amyotrophic lateral sclerosis and chronic spinal muscular atrophy. Charcot first taught that two distinct forms of

progressive amyotrophy may arise from lesion of motor cells: (1) protopathic, arising exclusively from the lesion in question; and (2) symptomatic, in which the nerve-cells are affected secondarily — consecutively, for instance, to a lesion of the white columns. This distinction was made as early as 1869.[27]

At a later date, Charcot emphasised this distinction still more strongly. He said that chronic protopathic spinal amyotrophy corresponded to the clinical type described by Cruveilhier, Duchenne and Aran. Here the spinal lesion is limited to the anterior cornua. On the other hand, chronic deuteropathic spinal amyotrophy has several varieties. The most important of these (amyotrophic lateral sclerosis) is characterised anatomically by symmetrical sclerosis of the lateral columns, with secondary damage to the cells of the anterior horns. Charcot admits, however, that symmetrical lateral sclerosis may occur without invasion of grey matter, and he says it is possible (though this is very doubtful) that a primary lesion of the grey matter may extend to the lateral columns.[28]

In 1885, the same observer again announced his belief in the existence of two forms of spinal muscular atrophy, both of them, of course, quite distinct from the primary myopathies which recent advances in medical science had shown to exist.[29]

Most modern writers have followed Charcot, but his theory has found two very powerful opponents in Dr. Gowers and Professor Leyden.[30]

Many more observations will be required before the point in dispute can be finally settled—namely, the point whether degeneration of the lower segment of the motor path ever occurs without coincident degeneration of the upper segment; or, in other words, whether every case which Charcot would have called protopathic spinal muscular atrophy is not, in fact, an example of what he would have called amyotrophic lateral sclerosis. The difficulty in coming to a decision is shown by the meagre results (so far as this question is concerned) of the careful investigation of a case recently published by

Dr. Mott,[31] and the actual truth must still be looked upon as unascertained; but there is no doubt that the evidence from autopsies, even in the earliest recorded cases, is very strongly in favour of Gowers's view.[32]

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[3] *Op. cit.* (third edition), 1844, *vide* p. 435 *et passim.*

[4] *Clinical Lectures on the Practice of Medicine* (second edition), 1848, vol. i, p. 509.

[5] *Arch. gén. de Méd.* (4e sér.), 1850, tom. xxiv.

[6] *Ibid.*, 1853, vol. i, p. 561 *et seq.* The paper was read to the Académie de Médecine in March, 1853.

[7] Obs. VI in Aran's collection of cases.

[8] *Bull. gén. de Thérap.*, 1853, vol. xliv, p. 295 *et seq.* The paper commences in the April number.

[9] *E.g.*, Gowers, *Diseases of the Nervous System* (second edition), vol. i, p. 471.

[10] *E.g.*, Putzel, *A Reference Handbook of the Medical Sciences* (New York), 1888, vol. vi, p. 569; Charcot, *Diseases of the Nervous System*, New Sydenham Society, 1877, p. 58, note.

[11] *Gaz. Méd de Paris*, 1860, p. 505.

[12] For Lockhart Clarke's three cases, see (1) Gairdner, Beale's *Archives of Medicine*, October, 1861, vol. iii; (2) Radcliffe, *British and Foreign Medico-Chirurgical Review*, July, 1862, vol. xxx, p. 215 *et seq.*; (3) Thudichum, Beale's *Archives of Medicine*, 1867, vol. iv.

[13] Duménil, *Gaz. Hebd.*, July, 1867.

[14] *E.g.*, Hammond, *Diseases of the Nervous System* (New York), 1871.

[15] Trousseau, *Clinical Lectures*, New Sydenham Society, 1868, vol. i, p. 126.

[16] Virchow's *Archiv*, 1855, Bd. viii, S. 537.

[17] Duchenne's Selected Works, New Sydenham Society, 1883.

[18] *Diseases of the Nervous System* (second series), New Sydenham Society, 1881, p. 176, note.

[19] "Progressive Muscular Dystrophy," *Clinical Lectures by German Authors*, New Sydenham Society, 1894, p. 234.

[20] "Ueber die Fortschreitende Bulbärparalyse und ihr Verhältniss zur Progressiven Muskelatrophie," Volkmann's *Sammlung Klinischer Vorträge*, 1873, No. 54. Translated by New Sydenham Society, in *German Clinical Lectures*, 1876.

[21] *Klinik der Rückenmarkskrankheiten*, 1875, Bd. ii.

[22] "Ueber die Hereditären Nervenkrankheiten," Volkmann's *Sammlung Klinischer Vorträge*, No. 171.

[23] *Deut. Arch. f. Klin. Med.*, 1884, No. 34. Erb states in this paper that he described and named the juvenile form in a work of his published in 1882.

[24] Selected Works of Duchenne, New Sydenham Society, 1883, p. 60.

[25] "De la Myopathie Atrophique Progressive," *Rev. de Méd.*, 1885 (February and April), tom. v. A *résumé* of this paper was presented to the Académie des Sciences in January, 1884. See also "Nouvelles Recherches," by the same authors, *Rev. de Méd.*, December, 1886, tom. vi.

[26] "Dystrophia Muscularis Progressiva," Volkmann's *Sammlung Klinischer Vorträge* (Neue Folge), May, 1890, No. 2. This has been translated by the New Sydenham Society for the third series of *Clinical Lectures by German Authors*, 1894.

[27] Charcot's *Lectures on Diseases of the Nervous System* (first series), New Sydenham Society, 1877, p. 59.

[28] *Diseases of the Nervous System* (second series), New Sydenham Society, 1881, p. 163 *et seq.*

[29] Lecture reported in a French journal in March, 1885. Translated by New Sydenham Society, 1889, in third series of Charcot's *Lectures on Diseases of the Nervous System.* Charcot gives here a classification of the chronic neuropathic and myopathic amyotrophies, but a more detailed one is given by Landouzy and Déjérine in their paper published in the *Rev. de Méd.*, 1885.

[30] See Gowers, *Diseases of the Nervous System* (second edition), 1892, vol. i, p. 472 *et seq.*

[31] *Brain*, Spring Number, 1895.

[32] See, *e.g.*, Kussmaul's Lecture on Bulbar Paralysis, *German Clinical Lectures*, New Sydenham Society, 1876, pp. 27, 30.

VI.

HISTORY OF BULBAR PARALYSIS.

VI.

HISTORY OF BULBAR PARALYSIS.

NOMENCLATURE.

Chronic bulbar paralysis.
Progressive muscular paralysis of the tongue, soft palate, and lips (*Duchenne*).
Progressive bulbar paralysis (*Wachsmuth*).
Glosso-labio-pharyngeal paralysis.
Bulbar nuclear paralysis (*Kussmaul*).
Progressive atrophic bulbar paralysis (*Leyden*).
Glosso-laryngeal paralysis (*Trousseau*).
Glosso-labio-laryngeal paralysis (*Duchenne*).

HISTORY.

The first published account of this disease in which it was recognised as a distinct affection appeared in 1860 in a paper by Duchenne, with the title "Paralysie musculaire progressive de la langue, du voile du palais, et des lèvres."[1]

In the preceding year, Duménil of Rouen had recorded a case of great importance from a historical point of view. It was one of progressive muscular atrophy of the usual kind, complicated, however, with non-atrophic paralysis of the muscles supplied by certain of the nerves arising from the medulla oblongata. At the *post-mortem*, the anterior spinal roots were found to be atrophied; but the bulbar nerves were also atrophied. Duménil therefore inferred that the cranial motor nerves have not the same influence as the spinal motor nerves over the nutrition of muscles.[2]

When Duchenne wrote in 1860, he said that his case was

something entirely different from progressive muscular atrophy. The latter, he said, was a lesion of muscular nutrition without paralysis; whereas the disease he was now describing was an instance of paralysis without atrophy, a condition which might remain isolated from beginning to end, though of course it might happen to be complicated by some other ailment, whether progressive muscular atrophy or anything else. As to the pathology of this new disease, the muscles of the tongue, palate, and lips were obviously at fault; but whether on account of some lesion of the central nervous system, or some neurosis, he was not prepared to say.

The first time Duchenne saw a case of bulbar paralysis was in 1852. When he published the second edition of his great work in 1861, he had collected fifteen cases. Twenty years before this (1841), Trousseau had met with a case of which he wrote out a description at the time; this was laid aside for long, but after Duchenne published his paper, it was brought to light again in connection with the famous "Clinical Lectures" at the Hôtel-Dieu. Apparently Trousseau did not see a second case till November, 1860.[3]

Accordingly, when Duchenne met with a case of combined muscular atrophy and bulbar paralysis, he thought that a mere chance had brought together two distinct diseases in one individual. Trousseau confirmed Duchenne's clinical observations, but inclined to the belief that the concurrence was due, not to a mere coincidence, but to an identity of anatomical lesion—viz., atrophy of motor roots. He did not yet recognise the affection as a central one.

The central character of the disease, and its connection with the medulla, were inferred, on theoretical grounds, by three German observers—viz., Baerwinkel of Leipsic (1860), Wachsmuth of Dorpat (1864), and Schultz of Vienna (1864). The first named shewed that the electrical reactions and other symptoms pointed to a central origin, while the distribution of the muscular affection corresponded to the medulla oblongata. Wachsmuth, in his important monograph, gave to the disease the now generally accepted designation, "Bulbar paralysis," and argued that the lesion must be

seated in the region of the bulbar nuclei described by Stilling.

Duchenne had, in 1860, merely hinted at the possibility of some central lesion, "anatomical or dynamic," affecting the origin of the bulbar nerves; but, as he himself says, he was here brought to a standstill until 1867, when the magnificent researches of Lockhart Clarke on the minute structure of the medulla threw a flood of new light on this obscure subject.

In 1869, Charcot and Joffroy published two cases of progressive muscular atrophy, in which the lips and tongue, besides the more commonly affected parts of the body, were involved. In each instance, they found, after death, atrophy of anterior spinal roots and of the hypoglossal and accessory nerves, with atrophy of the lateral columns, and extreme wasting of the ganglion-cells of the anterior cornua.

Leyden (1870 and later) published several cases of a similar kind, and mentioned disappearance of ganglion-cells in the hypoglossal nucleus.

Microscopic examination yielded confirmatory results in the case of Kussmaul and Maier in 1873.[4]

In 1870, Charcot expressed his belief that the morbid process attacks the motor nerve-cell in the first instance.[5]

As has been remarked, Trousseau believed that progressive muscular atrophy and bulbar paralysis are due to the same form of anatomical lesion. At the same time, he said that Duchenne was right in separating the two diseases, because the progress of the affection is different in the two cases. Kussmaul (1873) was perhaps the first who unreservedly urged[6] the identity of the two diseases, and no effectual attempt has been made since then to separate them.

Friedreich (1873) naturally opposed the neuropathic theory of bulbar paralysis. Duchenne and Joffroy (1870) had, as they believed, a formidable difficulty to deal with. They recognised in progressive muscular atrophy an atrophy without paralysis, and in glosso-laryngeal paralysis, a paralysis without atrophy. Yet in both cases the muscular affection was apparently dependent on changes in motor nerve-cells. They therefore argued that these nerve-cells must have two

important functions, the one trophic, the other motor; and they completed their theory by saying that in the one disease only the motor, and in the other only the trophic function of the cells is impaired. Friedreich condemned all this as an unwarrantable assumption, and said that in glosso-labio-laryngeal paralysis, as in progressive muscular atrophy, the changes in the nerve-cells are secondary to those in the muscles.[7]

He in his turn had to answer the question why progressive muscular atrophy and bulbar paralysis are so often met with together in the same patient. The reason, he said, is that the two are related as cause and effect. The more general muscular affection is the cause of bulbar paralysis. The connecting link between the two is an ascending neuritis. This begins in the nerves of the muscles involved in progressive muscular atrophy, and extends upwards till it reaches the cervical portion of the spinal cord. From this it readily passes to the medulla oblongata, and when it reaches the floor of the fourth ventricle, it can give rise to the symptoms of bulbar paralysis.

The question of muscular wasting in bulbar paralysis has been a bugbear to writers on the subject. It seems so contrary to what we should anticipate that there should not be wasting, and yet there was no feature of the disease on which Duchenne insisted more emphatically, and we know how careful he was to get his clinical information at first hand.[8] On the other hand, Kussmaul says the tongue is most commonly seen to be atrophied,[9] and Leyden speaks of the disease as characterised by muscular atrophy from the beginning.[10] As a matter of fact, it would appear that the most diverse conditions of the tongue have been met with in different cases. The tissue of the organ may be considerably altered in its characters without its bulk as a whole being appreciably diminished, and indeed without its naked-eye appearances being in any respect different from the normal. A remarkable case of this kind is recorded by Charcot.[11] Wasting, again, may be very considerable, or it may be absent. We should be prepared, on theoretical grounds, to meet with

instances of the last mentioned condition, because it is natural to expect that degeneration will sometimes occur in the upper segment only of the motor path between the cerebral cortex and those muscles which are innervated by the bulbar nerves —a change analogous to primary lateral sclerosis of the cord, in connection with which there is, of course, no muscular wasting.[12]

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[4] See Kussmaul on Bulbar Paralysis, *German Clinical Lectures*, New Sydenham Society, 1876.

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[6] Kussmaul, *loc. cit.*

[7] Friedreich, *Ueber Progressive Muskelatrophie*, &c., 1873.

[8] Selected Works of Duchenne, New Sydenham Society.

[9] Kussmaul, *loc. cit.*

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[11] *Lectures on Diseases of the Nervous System* (second series), New Sydenham Society, p. 374.

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VII.

HISTORY OF OPHTHALMOPLEGIA.

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HISTORY OF OPHTHALMOPLEGIA.

NOMENCLATURE.

Chronic, or progressive ophthalmoplegia.
Chronic nuclear ocular palsy.
Combined palsy of the ocular nerves.
Progressive palsy of the ocular muscles.
Progressive paralysis of the cerebral nerves.

HISTORY.

The term "ophthalmoplegia" is said to have been first used by Brümmer in 1850, to signify complete paralysis of the third nerve or pair of nerves. At a later date, Graefe used the expression "ophthalmoplegia progressiva," to indicate the condition now generally known by this name. In 1856, this author published a case of almost complete paralysis of the ocular muscles. Vision, however, and accommodation were intact; the pupil contracted during accommodation, but not on stimulation by light; the levator palpebræ acted perfectly on the right side, but imperfectly on the left side.[1]

Dr. Gowers states [2] that Graefe compared these ocular palsies with bulbar paralysis in 1868, and that Förster correctly localised the lesion, so far as concerns the external ocular muscles, in 1878.

Writing in 1873, Kussmaul stated that an attempt had been made, for instance by Benedikt, to extend the notion of bulbar paralysis, so as to include a progressive palsy of the cerebral nerves. As yet, however, no undoubted case of the kind had been recorded.[3]

In 1878, Mr. Jonathan Hutchinson proposed the designation "ophthalmoplegia interna" for a set of cases characterised by palsy of the internal muscles of the eye; palsy, that is, of the ciliary muscle, and of both circular and radiating fibres of the iris. Mr. Hutchinson had seen his first case in 1865. The facts of his eight cases suggested that the disease was nearly always due to syphilis, and that the lesion was probably in the lenticular ganglion, though conceivably it might be near the nucleus of the third nerve; but as the patients were all alive, there was as yet no *post-mortem* evidence to go upon.[4]

In the communication just referred to, Mr. Hutchinson proposed the name "ophthalmoplegia externa" for cases of symmetrical immobility of the eyes, with ptosis; that is, for cases of paralysis of the external ocular muscles. Mr. Hutchinson gave an account of fifteen cases of this kind in a paper published in the following year. He considered the initial lesion to be probably seated in the nuclei of the affected nerves, and to be, in most cases, a consequence of syphilis. The ocular palsies were occasionally associated with symptoms of locomotor ataxy, or with evidence of damage to the fifth, seventh, or eighth cranial nerve. No similar cases had as yet been recorded in England. The writer, who mentioned that Graefe recorded examples of this condition in 1867, and used the term "ophthalmoplegia" in connection with them, saw the first of his fifteen cases in 1862. His third case was under observation from beginning to end (1869 to 1876), and furnished the only autopsy. This patient always denied having had syphilis, and no signs or history of any syphilitic affection could be made out; moreover, two of his children whom he brought presented no suspicious sign. At last, however, in 1876, his eldest child, a girl aged twenty, was sent to Mr. Hutchinson with characteristic syphilitic keratitis, and she was then found to have notched teeth also.

When this man died, Dr. Gowers examined his brain and found in the nuclei of the third, fourth, and sixth nerves, changes similar to those which occur in the anterior cornua of the cord in progressive muscular atrophy.[5]

One of the earliest systematic writers to allude to ophthalmoplegia was Eulenburg of Greifswald, but there is practically nothing of historical importance in his chapter on this subject.[6]

Finally, reference ought to be made to an admirable address by Professor Ferrier on "The Pathology and Distribution of Atrophic Paralyses" (1893).[7]

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[2] *Diseases of the Nervous System* (second edition), vol. ii, p. 194, note.

[3] See Kussmaul on Bulbar Paralysis, *German Clinical Lectures*, New Sydenham Society, 1876, p. 42 *et seq.*

[4] *Medico-Chirurgical Transactions*, 1878, vol. lxi.

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VIII.

HISTORY OF THE PERONEAL TYPE OF MUSCULAR ATROPHY.

VIII.

HISTORY OF THE PERONEAL TYPE OF MUSCULAR ATROPHY.

NOMENCLATURE.

Peroneal type of progressive muscular atrophy (*Tooth*).
Peroneal type of family amyotrophy (*Gowers*).
Charcot-Marie type of progressive muscular atrophy.
Leg type of progressive muscular atrophy (*Sachs*).
Progressive neurotic muscular atrophy (*Hoffmann*).
Femoro-tibial type of Eichhorst (*Landouzy and Déjérine*).
Wetherbee ail.

HISTORY.

A short account may be given of the history of this disease, which still occupies a somewhat indeterminate nosographical position. It was first treated of as a distinct variety of muscular atrophy by Charcot and Marie in February, 1886.[1] These authors gave five cases of their own, and their paper contained abstracts of cases already recorded by other writers. Charcot and Marie thought that this disease was not a primary myopathy. Whether it was a myelopathy or multiple peripheral neuritis they considered doubtful; but they rather favoured the idea that it was myelopathic.

Later in the same year this affection was described (quite independently) by Dr. Howard Tooth, in his Cambridge Graduation Thesis, under the title of "The Peroneal Type of Progressive Muscular Atrophy," a designation which has come into very general use.[2]

A considerable number of cases of this affection had been described before 1886; but the peculiar features of the disease had, in some instances, not been fully appreciated. Among

those who published such cases were Eulenburg (1856), Eichhorst (1873), Friedreich, Oppenheimer, Ormerod[3] (1884), and Schultze (1884).

The "Wetherbee ail" probably corresponds most nearly to this malady. It is so called after a family named Wetherbee, many members of which were sufferers. Their pathological history was related by one of their number, E. H. Wetherbee, and was published by Hammond. E. H. Wetherbee and his father did not begin to suffer till they reached the age of thirty-nine; but the manner in which the disease affected the family generally makes it pretty evident that it was of the peroneal type. It began in the lower limbs, and fibrillary contractions often took place in the affected muscles.

An extensive family tree, showing many individuals affected by this disease, was published by Herringham in 1888.[4]

German neurologists declined at first, after Charcot, Marie, and Tooth published their researches, to admit an additional type of muscular atrophy; but ultimately Erb gave in his adhesion to the new views, as was shown by the communication published by his assistant Hoffmann in 1889.

The hitherto indeterminate pathology of this disease has been already adverted to. It will be sufficient here to mention some of the theories that have been held.

Schultze supposed his cases to be instances of multiple peripheral lesions. Landouzy and Déjérine, commenting on the observations of Eichhorst, Hammond, and Ormerod, expressed the opinion that the disease is a myopathy. Charcot and Marie thought it either multiple neuritic or spinal,—probably the latter. Tooth and Hoffmann think it a degenerative neuritis. Gowers thinks that, in different cases, nerves or muscles or both may be at fault. Sachs has suggested that this disease, as the "leg type of progressive muscular atrophy," is strictly comparable to the "arm type" (Duchenne's type), the difference being that hereditary influence is not so clearly proved in the arm type of Duchenne as in the other. Finally, Ferrier considers the peroneal type to be "primarily a hereditary myelopathy, occasionally complicated by neuritic changes, probably of a secondary character."[5]

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[2] See Tooth, *Brain*, July, 1887, vol. x, pp. 243-253.

[3] Ormerod, "Muscular Atrophy after Measles in Three Members of a Family," *Brain*, October, 1884, vol vii, p. 334.

[4] *Brain*, June, 1888, vol. xi.

[5] See Schultze, *Berl. Klin. Woch.*, 13th October, 1884, No. 41 ; Charcot et Marie, *loc. cit.* ; Gowers, *Diseases of the Nervous System* (second edition), vol. i, p. 534 ; Sachs, *Brain*, 1889-90, vol. xii ; Ferrier, *British Medical Journal*, 30th September, 1893.

IX.

HISTORY OF DISSEMINATED SCLEROSIS.

IX.

HISTORY OF DISSEMINATED SCLEROSIS.

NOMENCLATURE.

Disseminated sclerosis (sclérose en plaques disseminées).
Multiple sclerosis (with cerebrospinal, cerebral, and spinal varieties).
Insular sclerosis (*Moxon*).
Multilocular sclerosis.
Polynesic sclerosis.
Charcot's disease (*Althaus*).

HISTORY.

The history of this disease does not take long to write, partly because its evolution as a clinical and anatomical type is due largely to the labours of a very few individuals, and partly because, so far as our knowledge yet goes, its morbid anatomy and physiology are exceedingly simple when viewed alongside those of that highly complex entity, *tabes*, whose history is much older than that of modern neurology, and whose pathology still bulks so largely in the scientific study of chronic disease of the spinal cord.

It is generally stated by writers on the subject, that the lesions of multiple sclerosis were figured long ago by Cruveilhier and Carswell. Personally, I am not quite satisfied, after an examination of Cruveilhier's plates,[1] that the lesions he figures are those of this disease, although, when we remember the admirable field he had at his command, it seems exceedingly likely, *a priori*, that he *would* meet with genuine examples. One case figured by Carswell,[2] however, seems to be an undoubted instance.

Ludwig Türck (1855) recorded some examples, but he studied them only from the physiological point of view; so that apparently the disease had been lost sight of for a good many years.

Rokitansky, in his text-book, describes as being probably one of the results of encephalitis, a condition which corresponds fairly well to the lesion of disseminated sclerosis.[3]

In 1849, Frerichs recorded a case in which, for the first time, an earnest attempt was made to study the disease clinically, and to make a diagnosis during life.

In 1856, Valentiner published an article dealing with several instances of this disease, in which Frerichs, in his clinique at Breslau, had made a diagnosis during life.

In 1863, Rindfleisch of Zurich wrote a very important account of the histology of the disease. There is, he says, in the brain and cord, what Rokitansky would have called a focal increase of the interstitial tissue. Rindfleisch's work has not ceased to be of value. He states that the appearances point to the disease as starting in single blood-vessels with their branches. All the vessels in the diseased area, and also those in the immediate neighbourhood which still traverse healthy parenchyma, are in a condition which is characteristic of chronic inflammation. We must therefore, he says, distinguish three processes which run on in close association with one another: (1) changes in the vessels; (2) atrophy of the nervous elements; (3) metamorphosis of the connective tissue.[4]

Vulpian and Charcot recorded some fresh cases in 1862, and about this time the subject was taken up for thorough examination at the Salpêtrière. The results of Charcot's labours are embodied in a lecture delivered in 1868, which still constitutes—as regards typical cases at any rate—our standard description of the disease.[5]

Paralysis agitans was the disease with which disseminated sclerosis was particularly liable to be confounded. Charcot, in inspiring the thesis of his pupil Ordenstein (1867), endeavoured to make plain the difference between the two. In his lecture already referred to, Charcot alludes to a case reported by Baerwinkel some ten years before. The patient,

during life, was under Skoda's care, and the symptoms were carefully investigated and faithfully noted. In particular, it was observed that tremor showed itself only when voluntary movements were attempted. Paralysis agitans was the diagnosis arrived at, but after death, patches of sclerosis were found disseminated through all parts of the cerebro-spinal axis.

Again, a case recorded by Zenker, about 1865, was found at the autopsy to be one of multiple sclerosis. Yet Professor Hasse had made the diagnosis of paralysis agitans, though in the clinical notes emphasis was laid on the fact that tremor only showed itself on voluntary effort, or under the influence of emotion.

Charcot, lecturing in 1868, said that the disease was not yet known in England. At the Clinical Society of London, on the 9th April, 1875, Dr. Buzzard shewed a case of disseminated sclerosis, and a case of paralysis agitans (two diseases which Charcot had differentiated one from the other).[6] "Two cases of insular sclerosis of the brain and spinal cord" were recorded in papers published by Dr. Moxon in the same year.[7]

As has been already hinted, Charcot's description left but little to be added to a complete sketch of multiple sclerosis. It may be well, however, specially to state that Charcot first called attention to the persistence of the axis-cylinders—an observation which has been abundantly confirmed by later investigators. The same observer showed that the lesion is interstitial. Valentiner (1856) and Baerwinkel described isolated cases in which nystagmus was a symptom. Ordenstein (1867), Charcot, and Bourneville and Guérard (1869) shewed that nystagmus is a symptom of much importance in multiple sclerosis. Charcot found it present in about half the cases.[8]

The theory of Rindfleisch that the disease begins in the vessels did not at first meet with general acceptance, but there appears to be a disposition on the part of some writers nowadays to view it more favourably. Dr. Pierre Marie, accepting this doctrine, thinks that the disseminated sclerotic foci arise through embolisms, and he has, since 1884, urged

the importance of a preceding attack of some infectious disease in the etiology of multiple sclerosis.[9]

A word may be added here with regard to the nomenclature of this disease. "Inselförmige Sklerose," "insular sclerosis," "polynesic sclerosis," are among the designations employed for the general condition, and the foci of disease are spoken of as "islets" of sclerosis. Leube states[10] that Cruveilhier used the expression "îles" for the little grey patches of disease in the brain. The idea involved in the word "island" seems therefore to have commended itself to writers of various nationalities.

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